

# PRIMARY SYNOVIAL SARCOMA OF THE ABDOMINAL WALL: A CASE REPORT AND REVIEW OF THE LITERATURE

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إن الورم اللحمي المصلي هو ورم خبيث بالنسيج الأوسط ويشيع حدوثه في الأطراف لدى الكبار، وهو ذو صلة كبيرة بمحافظ المفاصل وأغلفة الوتد والأكياس المصلية وبنيات الأربطة. توجد حالات موثقة قليلة فقط لحدوث الورم اللحمي المصلي في الجدار البطن. في هذه الدراسة تعرض حالة للورم اللحمي المصلي الأولي الناشئ من سفاق الجدار البطني.

**الكلمات المرجعية:** الورم اللحمي المصلي ، أورام الجدار البطني.

*Synovial sarcoma is a malignant mesenchymal neoplasm which commonly occurs in the extremities of adults, in close association with joint capsules, tendon sheaths, bursae and fascial structures. Only a few cases of synovial sarcoma occurring in the abdominal wall have been reported. A case of a primary synovial sarcoma arising from the anterior abdominal wall fascial aponeurosis is presented.*

**Key Words:** Synovial sarcoma, abdominal wall tumor.

*J Fam Community Med 2008; 15(3):123-125.*

## CASE REPORT

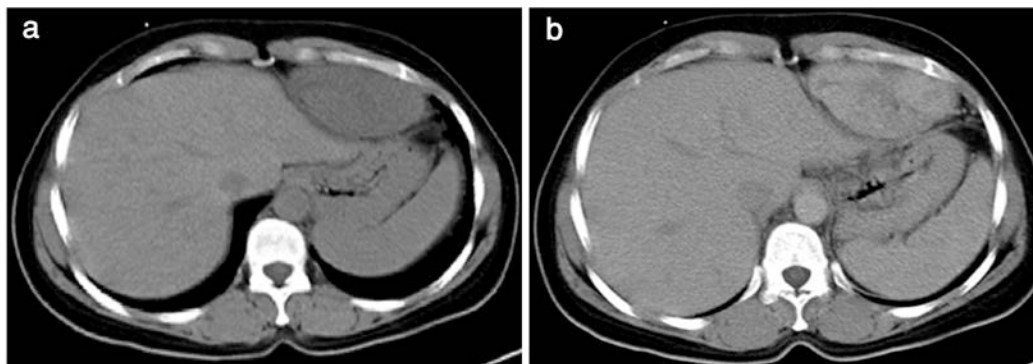
A 46-year-old married Saudi woman presented at a primary health care center with left upper abdominal pain, with no other associated symptoms. Her medical history was not remarkable. Based on the history and clinical examination, she was treated as gastritis. Five months later, she presented to the surgery clinic because of increased abdominal pain with no response to medication; her physical examination showed fullness of the left upper abdomen. Her routine laboratory investigation as well as chest and abdominal x-rays were not remarkable. Ultrasound of the abdomen showed a solid mass at the left hypochondrium separated from the spleen and just below the left hemidiaphragm. Computed Tomography (CT) scan of the abdomen (Figure 1) showed encapsulated hypodense mass at the left upper abdomen which was interposed but clearly separated from left liver lobe, spleen, stomach and left hemidiaphragm. Post intravenous contrast CT scan showed heterogeneous enhancement of the mass. Magnetic resonance

imaging (MRI) of the abdomen (Figure 2) confirmed the location of the mass and attachment to the inner aspect of the anterior abdominal wall but separated from internal abdominal organs. The mass was isosignal intensity to muscles in T1-weighted images and heterogeneous hyperintense signal in T2 weighted images. In post gadolinium T1 weighted images, the mass showed intense contrast enhancement.

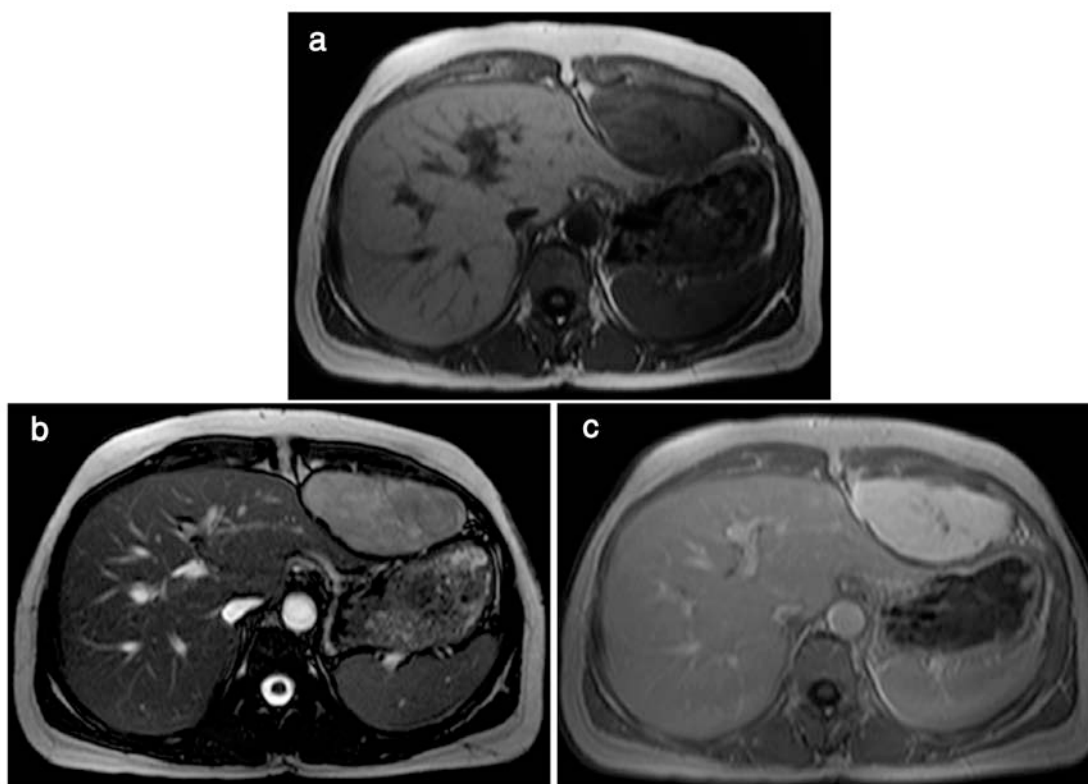
At surgery, a firm, rounded mass apparently encapsulated at the left sub-diaphragmatic area was found attached to the extra-peritoneal fascia of the anterior abdominal wall. Surgical excision was performed. Histopathology showed a biphasic tumor composed of spindle cell and epithelial cell elements, with mild anaplasia & infrequent mitosis. The immunostaining showed immuno activity for Vimentin. The final diagnosis was low grade synovial sarcoma involving the fascial aponeurosis of the anterior abdominal wall. The patient was referred to a specialized oncology center for follow-up.

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**Figure 1:** (a) Axial non-enhanced CT scan of the abdomen showing a well defined hypodense mass at the left hypochondrium, interposed between left liver lobe, left hemidiaphragm and stomach. (b) Contrast enhanced CT scan showing heterogeneous contrast enhancement of the mass.



**Figure 2:** (a) Axial T1W1: An oval shape mass with signal intensity parallel to the muscle. (b) Axial T2 W2: The mass is hypertense. (c) Axial post contrast T1W1 showing avid contrast enhancement of the mass.

## DISCUSSION

Synovial sarcoma (SS) is an uncommon soft-tissue malignant tumor that is common in the extremities of middle-aged patients, close to large joints particularly the knee in the popliteal fossa.<sup>1,2</sup> Despite its name, the lesion does not commonly arise in an intraarticular location but usually near joints. The tumor arises from pleuripotential mesenchyme, in close association with joint capsules, tendon sheaths, bursae and fascial structures. It is generally accepted that synovial sarcoma is derived from primitive mesenchymal

cells, not synovial cells.<sup>2</sup> Synovial sarcoma is the fourth most common type of soft tissue sarcoma following malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma. Synovial sarcoma accounts for 5-10% of all soft tissue sarcomas.<sup>3</sup> About 85-90% of SS occur in the extremities.<sup>4</sup> Reported examples arising in the anterior abdominal wall are rare. Synovial sarcoma in the abdominal wall tends to occur with a much greater frequency in females in contrast to such tumors in the extremities or the neck which

tend to occur with a much greater frequency in males.<sup>5</sup>

Radiological findings of SS are not pathognomonic. However, findings of a soft-tissue mass, particularly if calcified (30%), near but not in a joint of a young patient, are very suggestive of the diagnosis. Cross-sectional imaging features are essential for staging extent of the tumor and the planning of surgical resection.<sup>6</sup> The most common CT appearance of SS is that of a heterogeneous soft-tissue mass with attenuation similar to or slightly lower than that of muscle. CT is sensitive for the detection of calcification. Areas of lower attenuation representing necrosis or hemorrhage are also common with heterogeneous contrast enhancement.<sup>7</sup>

On MRI, SS typically appears as a prominently heterogeneous multilobulated soft-tissue mass with signal intensity similar to or slightly higher than that of muscle on T1-weighted MR images. Prominent heterogeneity with predominant high signal intensity is also a feature of SS on T2-weighted MR images. The presence of multilobulation and marked heterogeneity are highly suggestive of a diagnosis of SS. CT and MRI are useful in defining the extent of the disease and in follow up response to chemotherapy.<sup>7</sup> The presence of extensive calcification suggests a more favorable prognosis.<sup>8</sup> Similar CT and MRI appearances are seen in our case.

Despite its name, SS does not arise from synovium. There are three main histologic subtypes of SS: biphasic, monophasic, and poorly differentiated types. Monophasic SS predominantly consists of spindle cells resembling fibrosarcoma. Biphasic SS is typically composed of the spindle cell and epithelial cell elements. Poorly differentiated SS is generally epithelioid in morphology and has high mitotic activity.<sup>7,8</sup> In the present case, histopathology showed a biphasic pattern of SS, the coexistence of spindle cells in a wavy pattern, and the pseudoglandular formation. Both biphasic and monophasic synovial sarcomas are usually intermediate grade (grade 2/3); however, both types can be high grade (grade 3/3). Poorly differentiated synovial sarcomas are high-grade tumors.<sup>7</sup>

The specific chromosomal aberration in synovial sarcoma has recently been reported. The hallmarks for synovial sarcoma are the (X; 18) translocation and SYT-SSX gene fusion products.

This translocation is found in more than 90% of SS. Of current importance is the molecular diagnosis in synovial sarcoma, as well as immunohistochemical study, especially in cases in which histological diagnosis is difficult.<sup>7,9</sup>

Surgical excision is the treatment of choice, and the recurrence rate range from 28% to 36% even with adequate surgical and adjunctive therapies.<sup>4</sup> However, the multimodality treatment approach has improved the prognosis of synovial sarcomas. Tumor invasiveness, histologic grade and tumor size significantly correlate with the survival period, with a more preferable prognosis of synovial sarcomas encountered in childhood. The role of adjuvant therapy in the treatment of synovial sarcoma remains controversial. Chemotherapy has been used to treat metastatic or residual disease.<sup>7,10</sup>

Primary synovial sarcoma of the anterior abdominal wall is a rare extra-articular tumor site. In cases of anterior abdominal wall masses, it is important to consider the possibility of synovial sarcoma in the differential diagnosis.

## ACKNOWLEDGMENT

My sincere thanks to Dr. Fatma Almulhim, Professor of Radiology, and Dr Khaled Moghazy, Associate Professor in the College of Medicine at King Faisal University for their valuable support during preparation of this material.

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